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CONSULTANT INTERVENTIONAL CARDIOLOGIST

PROFILE INFO

Dr. Meet Thacker is presently associated with K. K. Patel Super Speciality Hospital, Bhuj as a Full Time Consultant Interventional Cardiologist .

He has rich experience of more than 5 years in the field of Non-invasive and Interventional Cardiology, Pediatric Cardiology and Structural Heart interventions.

PROFESSIONAL QUALIFICATION

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C.U. Shah Medical College,
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M.D. Medicine (Gold Medal)
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Sri Jayadeva Institute of Cardiovascular Sciences & Research, Bangalore

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Clinical Experience

- Dr. Meet is a young and Dynamic cardiologist having wide experience of more than 5 years in the Field of Non-Invasive and Clinical Cardiology, Interventional Cardiology including Pediatric and Structural Heart Interventions.
- He specializes in Primary Angioplasty in Myocardial Infarction (PAMI), Complex Coronary Interventions (CTO, Bifurcation PCI, LM Interventions), Coronary Imaging and Physiology guided Interventions, Carotid and Peripheral Vascular Interventions, Device closures of Congenital Heart Diseases like ASD, PDA etc.
- He Served as an Assistant Professor of Cardiology at Sri Jayadeva Institute of Cardiovascular Sciences & Research, from Sep-2020 to July-2021.
- He has worked as an Associate Interventional Cardiologist at Care Institute & Medical Sciences (CIMS Hospital) Ahmedabad with renowned Cardiologists Dr. Milan Chag and Dr. Keyur Parikh.

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Lance-Adams Syndrome: A Rare Case of Post-hypoxic Myoclonus, Developing After a Snake Bite

MEET M THACKER*, MANISH N MEHTA†

ABSTRACT

Lance-Adams syndrome (LAS) is a rare complication of a successful cardiopulmonary resuscitation (CPR) and is often accompanied by post-hypoxic action myoclonus. Less than 200 cases have been reported in medical literature till date. A 50-year-old female presented to the Emergency Department in a state of unconsciousness. Urgent intubation and CPR resulted in stabilization of vitals over a period of few days. On regaining consciousness, the patient developed myoclonus, which was characteristically present only on activity and absent at rest or during sleep. This action myoclonus was troublesome to the patient and interfering in the day-to-day activities of the patient. Patient was started on a combination therapy with levetiracetam and clonazepam, which resulted in marked diminution of myoclonus over a period of 15-20 days. Though LAS is a rare complication, proper diagnosis and prompt management may significantly reduce the morbidity and improves the quality-of-life.

Keywords: Lance-Adams syndrome, cardiopulmonary resuscitation, myoclonus, levetiracetam, clonazepam

Lance-Adams syndrome (LAS) is a rare complication of a successful cardiopulmonary resuscitation (CPR). LAS is known to present as action myoclonus, days to weeks after a successful CPR, due to hypoxic injury to brain. Post-hypoxic myoclonus (PHM) is divided into two types:

- The *acute type*, which is called "myoclonic status epilepticus," occurs within 12 hours in most cases after hypoxic brain damage in patients who are deeply comatose.
- The *chronic type*, called "the Lance-Adams syndrome," is characterized by action myoclonus beginning days to weeks after a successful CPR and persists in patients who have recovered consciousness after CPR.

LAS is a rare complication and less than 200 cases have been reported in medical literature till date. We present here a patient who was diagnosed as LAS after CPR due to cardiorespiratory arrest following a neurotoxic snake bite.

CASE REPORT

A 50-year-old female reported to our Emergency Department in a state of unconsciousness. Following a snake bite, she slipped into unconsciousness, as reported by the husband accompanying her. Her oxygen saturation on admission was 40% as measured by a finger oximeter, and respiratory movements were almost absent. After 10 minutes of vigorous CPR at the Emergency Department, her vital signs started to return. She was shifted to intensive care unit (ICU), and was given 20 vials of antisnake venom in total. She regained consciousness the next day, after being on intermittent positive pressure mechanical ventilation and vasopressor support for a day. Meanwhile, a computed tomography (CT) scan of the brain was done, which showed no significant abnormalities (Fig. 1). All routine investigations like complete blood count, liver function tests, renal function tests, urine routine and microbiological examination and serum electrolytes were within normal range as given in Table 1.

When she was tapered from sedation (midazolam) and muscle relaxation (atracurium), on the 3rd day of ICU, she developed a generalized seizure and subsequently myoclonic movements were continuously observed

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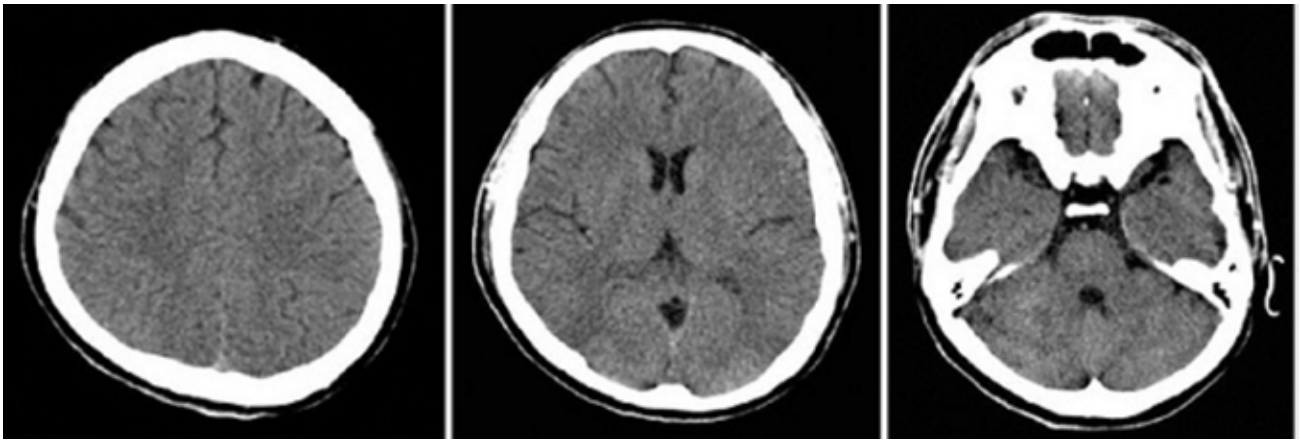

Figure 1. CT scan of the brain showing no significant abnormalities.

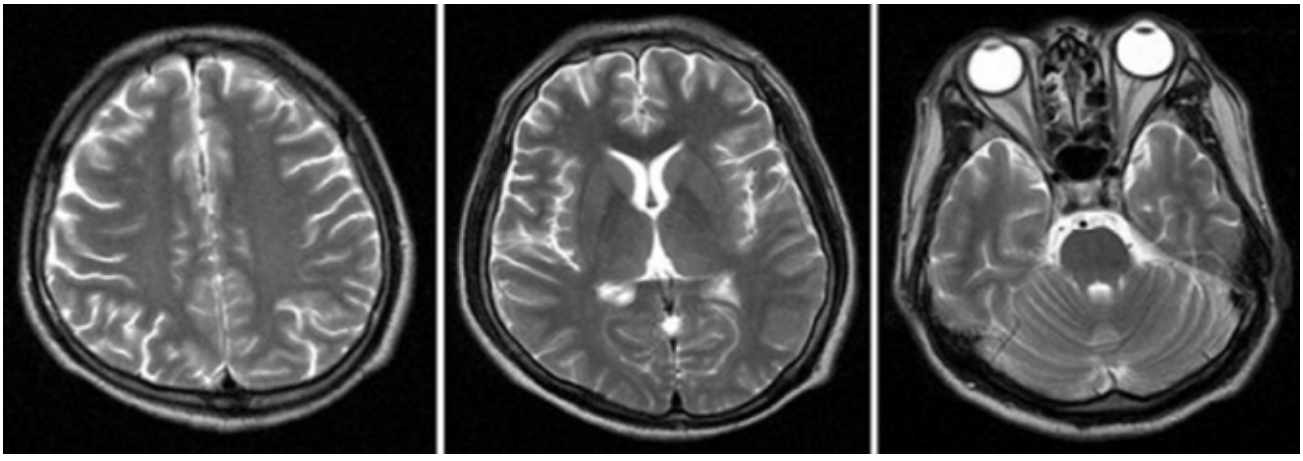

Figure 2. T_2-weighted MRI showing mild diffuse cerebral atrophy.

throughout her body, including face. At that time, the myoclonic movements were considered as generalized myoclonus secondary to hypoxic brain insult and were empirically treated with sodium valproate. However, these were not controlled with sodium valproate. The myoclonic jerks ceased with a single bolus dose of midazolam but the effect was transient. A repeat CT scan was meanwhile done, which showed no abnormalities and the T_2-weighted magnetic resonance imaging (MRI) showed mild diffuse cerebral atrophy (Fig. 2). On Day 5, the patient's mental status improved, she was now intermittently able to obey simple commands, but the myoclonic jerks continued. The rest of the central nervous system (CNS) examination was normal except higher functions, where the patient had dysarthria, dyscalculia and some attention deficits.

Subsequently, on Day 9, the patient was shifted to ward and started on levetiracetam (500 mg b.i.d.) and subsequently clonazepam (1 mg t.d.s.) was added. With these medications and physiotherapy, the patient started showing improvement, in the form of reduced frequency of myoclonic jerks, and also some improvements in higher functions. Patient was discharged on Day 21 when

Table 1. Routine Investigations

Test	Value
Hemoglobin	10.8 g/dL
Total WBC count	15,200 cells/mm^3
Differential count (N/L/M/E/B)	66/20/12/02/00%
Platelet count	1,64,000 cells/mm^3
PT/INR	14.4 sec/1.1
Serum bilirubin (total/direct/indirect)	1.2/0.4/0.8 mg/dL
SGPT	32 IU/L
Serum creatinine	0.8 mg/dL
Blood urea	28 mg/dL
Serum Na^+	138 mEq/L
Serum K^+	4.6 mEq/L

Mauriac Syndrome Presenting as Primary Amenorrhea in a Case of Type 1 Diabetes Mellitus

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ABSTRACT

Type 1 diabetes mellitus (T1DM) is a common occurrence and Mauriac syndrome is a well-documented complication of poorly controlled T1DM. It usually presents with hepatomegaly due to hepatic glycogen deposition, pubertal and growth delay, hypogonadism, dyslipidemia, protuberant abdomen, cushingoid features and elevated transaminases. We present here a case of an adolescent female with T1DM who presented with primary amenorrhea with cushingoid facies and poorly controlled diabetes taking premix insulin. With adequate glycemic control, the metabolic abnormalities were reversed and she had menarche at the age of 21 years.

Keywords: Diabetes mellitus, type 1 diabetes, complications of diabetes, Mauriac syndrome

Poorly controlled diabetes is a frequent problem in developing countries like India, leading to many complications related to inadequate glycemic control and Mauriac syndrome is one of them. Mauriac syndrome complicates type 1 diabetes mellitus (T1DM) and includes short stature, glycogen laden enlarged liver leading to hepatomegaly, dyslipidemia, growth maturation and pubertal delay, moon facies, protuberant abdomen and proximal muscle wasting and it is also frequently associated with other microvascular complications like retinopathy and nephropathy.[1,2]

Commonly seen in children, adolescent and teens, it occurs equally in both sexes, and usually is seen in patients on plain insulin or premix insulin and with poor control of diabetes mellitus. With adequate control of blood sugar and with the advent of longer acting basal insulin, the incidence of this syndrome has been decreasing in the present era. With good glycemic control, most of the manifestations of this syndrome can be reversed; thus making a prompt diagnosis and effective and timely intervention is important.[3]

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CASE REPORT

Ms S, a 19-year-old unmarried female was admitted in our ward with poorly controlled diabetes. A known case of T1DM since the age of 8 years, she was in follow-up of Pediatrics Department till 15 years of age and had no medical contact for the last 3 years. She was taking premix insulin (30:70 combination of regular insulin and NPH insulin) in the dose of 20 units in the morning and 12 units at night for the last 3 years. Upon admission, patient had a blood sugar (random) of 450 g/dL and urine had +3 sugar with no ketones by dipstick method.

She denied any history of fever, nausea, vomiting, abdominal pain, burning micturition, etc. On general examination, she had stunted growth with a height of 134 cm (<95 percentile) and weight of 42 kg.

General examination was also significant for 'moon-like' face (Fig. 1), protuberant, globular abdomen and less developed secondary sexual characteristics like sparse axillary and pubic hair. Breast development was Tanner's stage II. Upon enquiry, she gave a history of primary amenorrhea. Blood and urine investigations on admission showed serum creatinine of 1.2 mg/dL, blood urea of 32 mg/dL, hemoglobin of 8.4 g/dL, total count of 11,400/μL. Urine sugar was +3 and urinary ketones absent by dipstick; 24 hours urinary protein excretion was 492 mg/24 hours. She had a glycosylated hemoglobin (HbA1c) of 12.2% suggesting markedly impaired glycemic control. Her thyroid function tests were within normal range. Lipid profile showed a total

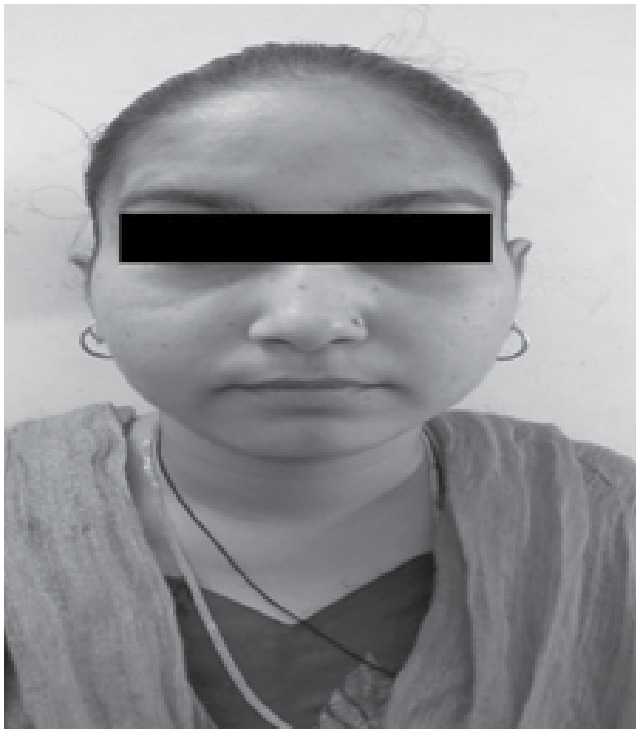

Figure 1. Characteristic moon-like face.

cholesterol of 320 mg/dL, low-density lipoprotein (LDL) was 170 mg/dL and triglycerides were 270 mg/dL.

Subsequent evaluation by ultrasound of abdomen revealed hepatomegaly, hypoplastic uterus and evidence of renal parenchymal disease (loss of corticomedullary differentiation in bilateral kidneys). Fundoscopy revealed Grade 2 nonproliferative diabetic retinopathy. Thus, she was a case of poorly controlled T1DM, with nephropathy, retinopathy, hepatomegaly, characteristic moon-like face and primary amenorrhea. Liver biopsy was also done, which showed characteristic glycogen deposition and steatosis (Fig. 2), confirming the diagnosis of Mauriac syndrome.

The patient was subsequently put on a basal-bolus regimen with insulin glargine at bedtime and three doses of plain insulin before three major meals. She was also counseled about adequate dietary restriction and a strict compliance was ensured.

After 6 months of strict adherence to basal-bolus regimen and ensuring dietary and drug compliance, her HbA1c fell to 8.0% and thereafter was constantly maintained around 7-7.5%. With this strict control, after 2 years, she had her menarche at the age of 21 years and also showed improvement of growth as well as regression of hepatomegaly and other metabolic abnormalities including dyslipidemia.

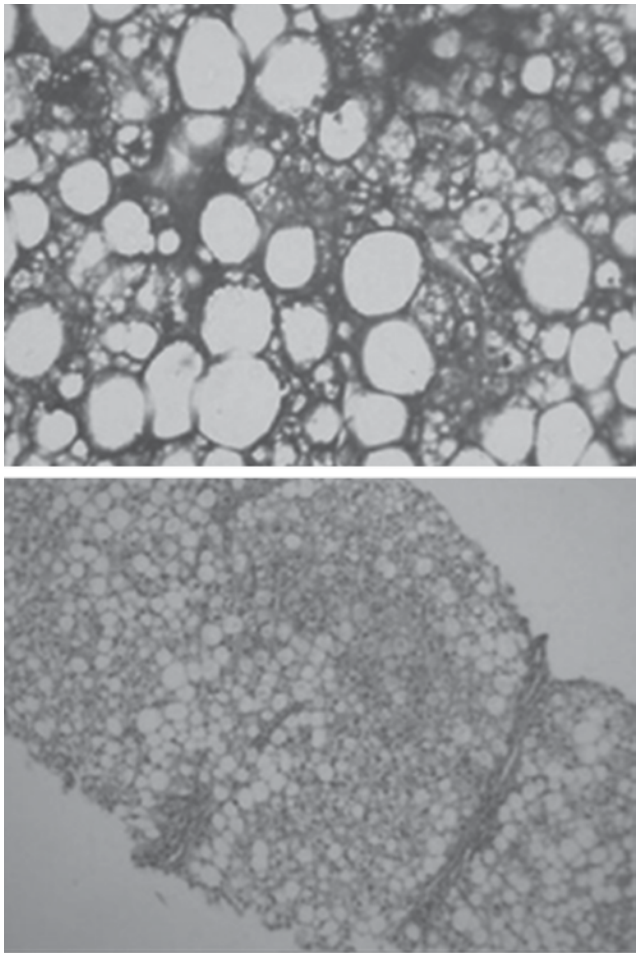

Figure 2. H&E stained liver biopsy at high and low magnification.

DISCUSSION

Mauriac syndrome was first described by Mauriac in 1930 in children with T1DM presenting with clinical features of growth failure, maturation delay, hepatomegaly and abdominal distension.[4] This syndrome is related to poorly controlled insulin-dependent diabetes of long duration. With improved patient care and the use of longer acting basal insulin, the incidence of this syndrome has become unusual and severe growth failure or pubertal delay is rare in juvenile diabetic patients.[5,6]

Other clinical features of this syndrome consist of dyslipidemia, cushingoid facies, elevated liver transaminases, glycogen deposition in the liver and delayed maturation. The pathogenesis of Mauriac syndrome is not clear but thought to be multifactorial. The features of Mauriac syndrome are mostly related to fluctuating levels of glucose and insulin with both

periods of underinsulinization and overinsulinization contributing to the presentation. Inadequate glucose to the tissues, decreased insulin-like growth factor 1 (IGF-1), growth hormone (GH) levels, hypercortisolism and resistant or defective hormone receptor action contribute to stunted growth and delayed puberty. The periods of supraphysiological levels of insulin are associated with glycogen deposition in the liver leading to hepatomegaly.[7,8]

Blood glucose passively enters the hepatocytes in which glycogen synthesis is promoted by high cytoplasmic glucose concentration reliant on the presence of insulin. Glycogen is then trapped within the hepatocytes as a result of a vicious cycle of hyperglycemia and insulin treatment.[9] Poor glycemic control due to hypoinsulinemia leads to lipolysis and ketone liberation. Ketosis activates cortisol synthesis promoting the release of fatty acids and hyperglycemia.[10]

Liver biopsy is helpful to confirm the diagnosis of Mauriac syndrome. Histologic features are characterized by large, swollen, glycogen-laden hepatocytes and glycogenated nuclei without significant fatty change, inflammation, lobular spotty necrosis or fibrosis.[9]

A high index of suspicion is required for the diagnosis of this syndrome. Tight glycemic control, preferably by basal-bolus insulin regimen, usually leads to improvement in hepatomegaly and dyslipidemia, and majority of the metabolic consequences may be reversed, with improvement in growth and pubertal changes.

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Serum Iron and Ferritin Levels Associated with Microvascular Complications in Type 2 Diabetes

Raised iron and serum ferritin levels have a significant positive association with HbA1c levels in patients with poorly controlled type 2 diabetes, says a new study presented at the American Association of Clinical Endocrinologists (AACE) annual meeting in Austin, Texas. These associations were also observed for microvascular complications of type 2 diabetes, including nephropathy, retinopathy and neuropathy.

Adrenalectomy Effective for Young Female Patients with Unilateral Primary Aldosteronism

Adrenalectomy can be effective treatment for unilateral primary aldosteronism, particularly in younger and female patients, suggests the Primary Aldosteronism Surgical Outcome (PASO) study reported online May 30, 2017 in *The Lancet Diabetes and Endocrinology*. Fewer antihypertensive medications and absence of left ventricular hypertrophy were other factors independently associated with complete clinical success. Researchers recommend evaluation of outcomes in the first 3 months post-surgery, again at 6-12 months and then annually.